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Diffuse large B-cell lymphoma of non-germinal center type of the buttock

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Abstract Primary lymphoma of soft tissue is very rare. An 80-year-old man presented with a right buttock tumor measuring 5 × 5 × 4 cm. Tumorectomy was performed. The tumor was located in the fascia and subcutaneous tissue, and encapsulated by a thin capsule. Histologically, the tumor showed atypical lymphoid cell proliferation. Immunohistochemically, the tumor cells were positive for CD45, CD20, CD79a, and CD138. The Ki-67 labeling was 90%. In contrast, they were negative for pancytokeratins, epithelial membrane antigen, CD10, MUM1, bcl-6, cyclin D1, CD3, CD30, CD45RO, TdT, CD56, S100 protein, HMB45, chromogranin, α-smooth muscle actin, and desmin. A diagnosis of diffuse large B-cell lymphoma (DLBCL) of soft tissue was made. The subtype of DLBCL is non-germinal center type (non-GC). Systemic investigation using various imaging modalities including CT, MRI and Ga-scintigraphy revealed small nodular lesions up to 1 mm in the lungs and liver. However, biopsies of these locations were not performed because of his old age. The nodular lesions of the liver and lungs were small in diameter (up to 1 cm), and it is strongly likely that the buttock tumor is primary. The patient was treated by R-CHOP chemotherapy and radiation. He was referred to another specialized hospital for his will.
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1. Introduction

Many kinds of malignant tumors occur in the soft tissue. Malignant lymphoma of the soft tissue is extremely rare. For example, Travis et al showed that among 7000 malignant lymphomas, only 8 (0.11%) were soft tissue malignant lymphomas [1]. In the literature, several case reports and case series of soft tissue malignant lymphoma have been reported [1–9].

Diffuse large B-cell lymphoma is characterized by high-grade lymphoma exhibiting large nuclei and B-cell markers such as CD45, CD79a and CD138. Exclusion of other type of B-cell neoplasms is necessary in making the diagnosis of DLBCL. DLBCL is now classified into 2 types: DLBCL of germinal center B-cell (LBCL GCB) and DLBCL of non-germinal center B-cell (DLBCL-non-GCB) [7] types. Recent advance in antibody cancer therapy has improved the outcome of B-cell neoplasms. The anti-CD20 antibody drug (rituximab) is powerful in the treatment of B-cell neoplasms. Now, the 5 year and 10 year survival of DLBCL is circa 70% and 50% [7]. The prognosis is better in DLBCL in GCB type than in DLBCL in non-GCB type [7]. The author herein reports a case of DLBCL non-GCB occurring in soft tissue

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(buttock). Current WHO classification of lymphomas was employed [9].

2. Case report

An 80-year-old Japanese man consulted to our hospital because of right buttock tumor measuring 5 × 5 × 4 cm. Abnormal blood date included mild diabetes (HBA1C; 7.1%), mild hypercholesterolemia, and mild elevation of liver enzymes. The tumor markers were within normal ranges. The tumor was hard, and overlying skin was free from tumor involvement. CT also detected a solid tumor in the deep soft tissue of buttock. The tumor had ragged border and no cystic areas were seen in the CT images. Tumorectomy was performed. The tumor was located in the fascia and subcutaneous tissue, and encapsulated by a then capsule.

Histologically, the tumor showed atypical lymphoid cell proliferation (Fig. 1A). No follicular structures were recognized. The atypical cells invaded the surrounding fat tissue. Higher power view showed that the atypical round cells are large lymphoid cells with hyperchromatic nuclei and nucleoli (Fig. 1B). Mitotic figures and apoptotic cells were scattered. Features of Burkitt lymphoma were not seen. Plasma cells were not prominent.

An immunohistochemical study was performed with the use of Dako Envision method and its variation, as previously described [10–13]. Immunohistochemically, the tumor cells were positive for CD45, and CD20 (Fig. 1C), CD79a, and CD138. The Ki-67 labeling was 90% (Fig. 1D). In contrast, they were negative for pancytokeratins, epithelial membrane antigen, CD10, bcl-6, MUM1, cyclin D1, CD3, CD30, CD45RO, TdT, CD56, S100 protein, HMB45, chromogranin, α-smooth muscle actin, and desmin. A diagnosis of diffuse large B-cell lymphoma (DLBCL) of the soft tissue was made. The subtype of DLBCL was non-germinal center B-type (non-GCB). (CD10−, bcl-6−, MUM1−).

Systemic investigation using various imaging modalities including CT, MRI and Ga-scintigraphy revealed small nodular lesions in the lungs and liver. However, biopsies of these locations were not performed because of his old age. The patient was treated by R-CHOP chemotherapy and radiation. He was referred to another specialized hospital for his will. The nodular lesions of the liver and lungs were small in diameter (up to 1 cm), and it is strongly likely that the buttock tumor is primary.

3. Discussion

The present tumor showed diffuse proliferation of large lymphoid cells. The histological, cytological, and immunohistochemical features are indicative of high grade B-cell neoplasm (DLBCL) [9]. High Ki-67 labeling (90%) indicates malignant nature of the tumor. The present case is obviously not low grade B-cell lymphomas (prominent features: small lymphoid cells), such as small lymphocytic lymphoma/CLL (prominent features: monotonous small lymphoid cells), follicular lymphoma (prominent features: follicular features and positive bcl-2), mantle cell lymphoma (prominent

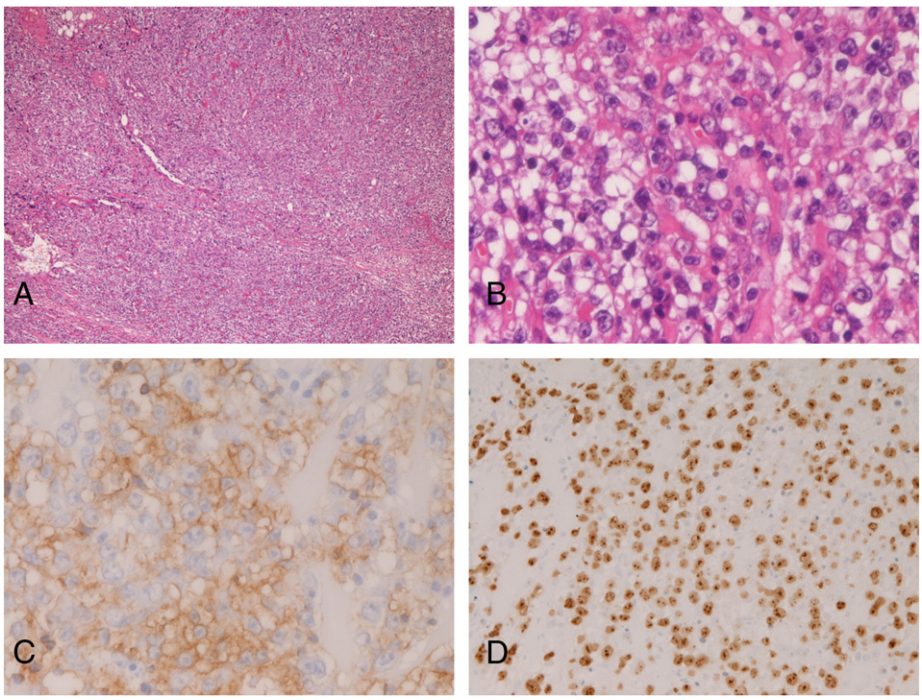

Fig. 1 Histological and immunohistochemical features. A: Diffuse atypical lymphoid cell proliferation is seen in the subcutaneous fat tissue. HE, ×400. B: The atypical cells have hyperchromatic nuclei and occasional nucleoli. Some mitotic figures and apoptotic cells are seen. HE, ×400. C: The tumor cells are positive for CD20. Immunostaining, ×400. D: The Ki-67 labeling is 90%. Immunostaining. ×400.

features: characteristic tumor small lymphoid cells and positive cyclin D1), lymphoplasmacytic lymphoma (characteristic features: small lymphoid and plasma cell proliferation positive for CD138), and MALT lymphoma (characteristic features: follicular formation, lymphoepithelial lesions, monoxytoid B-cells, follicular colonization, and centrocyte-like cells) [8]. The present case is not Burkitt lymphoma; the present tumor lacked features of Burkitt lymphoma such as proliferation of immunoblastoid cells and starry-sky appearances. The present case is not immunoblastic lymphoma histologically (lacks immunoblastic features such as prominent central-located large nucleoli), and also because of negative CD30 and TdT [9].

Soft tissue lymphoma can develop in any sites, but are common in the extremities [3]. Soft tissue lymphoma can occur at the sites of previous surgery [5]. Travis et al. [1], who examined 8 cases of soft tissue lymphoma, stated the locations of thigh (four), upper arm (two), ankle (one) and calf (one). In the 19 cases of Salamao et al. [4], the locations were lower extremity (seven), upper extremity (six), chest (three), gluteal region (two) and frontal subgaleal region (one). In a large series by Lanham et al. [3], the locations of soft tissue lymphoma were as follows: thigh in 25, trunk in 22, leg in 8, forearm in 4, buttock in 4, and foot in 1.The present tumor developed in the left leg. Most cases of soft tissue lymphoma are of B-cell type [1,3–6], while T-cell lymphoma is reported [2]. In the series of Salamao et al. [4], of the 19 soft tissue lymphomas, 11 were DLBCLs, 7 were low grade B-cell lymphomas, and 1 was peripheral T-cell lymphoma. In the 8 cases of Travis et al. [1], 4 were low grade B-cell lymphomas, 3 were DLBCLs, and 1 was lymphoblastic lymphoma. The diagnosis is usually biopsy or tumorectomy [1–5], but fine needle aspiration is also useful [6]. The treatment is usually chemotherapy and radiation [1–6]. The prognosis depends on tumor stage and histological type [1–6]. In DLBCL, R-CHOP with radiation is the first choice, and has a nice effect with approximately 40% survival in 10 years [7]. Although there are no significant data on the outcome in soft tissue lymphoma, it seems that soft tissue DLBCL shows poorer prognosis than nodal DLBCL [8].

The present case was associated with nodular lesions of the lungs and liver. Although biopsies were not performed, these lesions may be lymphoma involvements or another tumor. The nodular lesions of the liver and lungs were small in diameter (up to 1 cm), and it is strongly likely that the buttock tumor (5 × 5 × 4 cm) is primary.

Finally, although imaging findings of CT and MRI are out of the scope of this report, there are some references of the imaging features of soft tissue lymphomas [14]. The pathogenesis of soft tissue lymphoma is not known, but it seems that chromosomal abnormality, abnormality of oncogenes and anti-oncogenes, abnormal, and abnormal metagenetic alteration may play a role in the oncogenesis. Carcinogens are known to be responsive for the carcinogenesis in some cases of soft tissue lymphoma [15,16].

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